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The Straight Poop on Constipation and Its Treatment

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One of the most frequent referrals to the pediatric gastroenterologist is for the evaluation and management of constipation. It is a very common concern among parents, children, and providers alike and presents extraordinarily frequently to primary care practitioners caring for children of all ages. In the vast majority of instances, the nature of the constipation is functional and never requires referral to a sub-specialist. In this article, I hope to briefly review the pathophysiology of functional constipation and how to distinguish this from pathological forms of constipation. I would also like to propose some general guidelines for the management of this disorder and to point out some of the common pitfalls encountered when trying to do so. My goal here is to

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In this edition

Imaging of the Child with Intussusception

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A recent controversy at Wilford Hall in the imaging work-up of intussusception made me think of a recent personal anecdotal experience. During an overseas phone conversation in Turkey, this past February, my parents informed me that my 8-month-old nephew was sick. Apparently he had been vomiting and there had been some blood in his stools. I immediately called my sister and upon further investigating, discovered that his vomit was bright green and yellow and that he was somewhat lethargic. She said that she had been very concerned until a “medical friend” had told her that her son “just has a bug” and would be fine. After talking to her she went immediately to the ER where an indeterminate plain film was obtained. With high clinical suspicion, a therapeutic/diagnostic enema was then performed and the intussusception was successfully reduced.

In the imaging work-up of intussusception a plain film is obtained to exclude free air, and to look for signs of intussusception. Plain film signs of intussusception, from most to least sensitive are as follows: 1. Soft tissue mass in the right upper quadrant with possible air crescent and target sign. 2. Small bowel obstruction. 3. Lateralization of the ileum. 4. Paucity of gas in the right lower quadrant. 5. Absence of stool in the cecum. . A review of the literature for the plain film diagnosis of intussusception shows a sensitivity of 80-90% and a

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Got Rash?

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Few things are as disturbing to parents as an itchy, febrile, splotchy child. “What is it? What should we do? Is it contagious?” are questions brought up by the anxious parent who needs to get back to work. Pediatric exanthems are a collection of host responses which may be specific enough to diagnose the offending agent or non-specific to the point that even the dermatologist will sign off the case as “viral exanthem”. What follows are descriptions of several entities which occur frequently and several which are rare in the

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specificity of 58-90%. The plain film findings are categorized as positive, negative, or indeterminate (2).

In the result of a positive plain film, no diagnostic studies are needed. The pediatric surgeon should be notified/consulted and a therapeutic enema should be performed immediately.

An indeterminate exam requires further diagnostic investigation. Ultrasound has proven to be 100% sensitive and specific in the diagnosis of intussusception, in the hands of an experienced pediatric radiologist (1). With ultrasound there is no radiation to the patient and the discomfort of the enema study is avoided. Ultrasound also offers the ability to diagnose possible abnormalities that may mimic intussusception such as neoplasms, congenital cysts or bowel wall thickening (various causes). If an experienced radiologist is not available, a diagnostic enema may be performed to exclude the diagnosis.

A negative plain film may be problematic and requires the oft-quoted phrase, "Clinical correlation is recommended." In a large study published in Pediatric Radiology the diagnostic accuracy of abdominal x-rays when read as negative was 95% (2). The idea is that high clinical suspicion (by an experienced pediatrician) for intussusception is enough to require further diagnostic evaluation even if plain films are read as negative. If clinical suspicion is low or intermediate then no further imaging is required.

Other recent issues in the imaging of intussusception deal with predicting reducibility by ultrasound and using ultrasound guidance for both water and air reductions. Findings by ultrasound that resulted in a high success rate (100%) included: the target sign, and an outer (intussuscipiens) bowel wall thickness of less than 7mm. The reducibility percentage decreases with increasing outer bowel wall thickness and is dramatically reduced when a small amount of fluid is seen around the intussusceptum. Free fluid did not seem to affect the reducibility of the intussusception (3).

Most would consider ultrasound as a non-invasive diagnostic modality, however with the introduction of ultrasound contrast many procedures can now be performed under ultrasound guidance (4). Hydrostatic ultrasound guided reductions of intussusception is becoming more wide spread. The disadvantage is that with air reductions, should a perforation occur, it is typically a micro perforation that may be difficult for the surgeon to find. With hydrostatic perforations the perforation is typically much larger. In May of 2000, Gu et al, published their research showing that ultrasound can be used to guide air reductions. The method is more time consuming as repeat ultrasounds are often required to confirm reduction up to an hour after therapy, however, this would eliminate the need for any radiation exposure (5).

Whatever the method of diagnosing a child with intussusception and reducing it, the role of the clinician remains vital. Providing the clinical suspicion of intussusception in the history will likely increase sensitivity of the radiologist to the diagnosis and will result in better and more expedient care of the patient.

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The Multicystic Dysplastic Kidney

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The multicystic dysplastic kidney is the most common form of renal cystic disease in childhood, and one of the most common causes of abdominal masses in neonates. Its frequency is equal to that of hydronephrosis. Sixty-five percent of these kidneys are diagnosed by prenatal ultrasound, and 91% are diagnosed within the first year of life. The incidence is from 1/1000 to 1/3400 live births. The MDK occurs more frequently on the left than on the right. Some sources report that the disorder is more frequent in girls, some say that it is more common in boys and some say that the incidence is equal in the sexes. (It really does not matter, anyhow, does it?)

MDK is unilateral in 80-90% of cases; bilateral disease is not compatible with life. Infants with bilateral disease die from the respiratory consequences of oligohydrdaminos.

The renal parenchyma of the multicystic dysplastic kidney is completely replaced by multiple cysts held together by connective tissue with no normal renal tissue noticeable. The appearance has been compared to that of a "bunch of grapes". The cysts vary in size but most of them are large and they usually do not communicate with each other. Most MDK are associated with absence or atresia of the ureter and absence of the renal pelvis. Histologically the tissue consists of undifferentiated ducts and glomeruli. Primitive ducts are necessary for the diagnosis.

The contralateral kidney is abnormal in up to 40 % of patients with MDK. The most common finding in the other kidney is hydronephrosis secondary to UPJ obstruction. Next in frequency are ectopic ureters, duplicated ureters, and VUR. Abnormalities of other organs may also be associated with MDK, one of the more frequent being esophageal atresia with TE fistula. There is an increased incidence of MKD in VATER association and branchio-oto-renal syndrome. Most cases are sporadic, having no hereditary pattern.

The primary tool for diagnosing MDK is renal ultrasound. The prenatal diagnosis can be made between the 21st and 35th weeks of gestation – the average time of diagnosis is 28 weeks. In utero and postnatally, it may be difficult to distinguish MDK from severe hydronephrosis. The MDK has no functioning tissue but the hydronephrotic kidney usually has a rim of viable parenchyma. A nuclear DMSA is helpful in distinguishing between the two. There will be no uptake of the nucleotide in the MDK.

The clinical presentation is usually that of a left-sided, freely movable, non-tender, palpable mass in an asymptomatic newborn. The mass will sometimes transilluminate. Occasionally the mass may be large enough to crowd the abdominal space and induce abdominal distention, nausea, vomiting, and shortness of breath. Very rarely will the infant have hypertension. The urinalysis is usually normal. Adults may present with proteinuria, hematuria, infection, hypertension, and abdominal pain. The MDK may appear calcified on ultrasound in adults.

All infants suspected of having a MDK must have a VCUG and a DMSA study. It is important to distinguish between MDK and hydronephrosis because the follow-up differs. There continues to be great controversy about the need for nephrectomy in children with MDK. There is a very small but real potential for malignant degeneration. Wilms tumors and renal cell carcinomas have developed in these kidneys. If the kidneys are left in place, intensive follow-up is needed during the first few years and periodic follow-up for life. A renal ultrasound should be done every 3-6 months for the first two years, then every 6 months until the child is 5 years old and annually thereafter.

On the other hand, the pendulum may begin to swing back in the direction of early surgical removal of MDK. There is a tremendous commitment on the part of the physician and the patient, plus a significant cost involved in the life-long ultrasound follow-up of patients with MDK. In addition, even though the cystic changes as seen on ultrasound may disappear, there may still be solid elements left in which a tumor could grow. Removing the MDK surgically can be done as an outpatient, with the whole procedure taking about one hour. The surgical risks are low, and the patient would be spared the worry about the complications associated with the presence of a multicystic dysplastic kidney.

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immunized, industrial setting but, when they occur in the US or are seen on humanitarian assistance missions abroad, should be recognized because of their medical significance.

MEASLES
(Rubeola)

The etiologic agent is a paramyxovirus in the genus Morbillivirus and is characterized by rash mimicked by many other conditions. Called "morbilli" to distinguish from "morbus" (plague) the term "morbilliform" is now firmly in the dermatological lexicon although measles itself is now quite rare in the United States. It is spread by direct contact with infectious droplets or by airborne spread. Winter and spring are the most likely times of year to see measles. Incubation of this virus from exposure to onset of symptoms is 8 to 12 days. During the incubation period, virus may be cultured from the mucosa of asymptomatic individuals

A symptomatic prodrome phase (about 4 days) ensues with fever, malaise, cough, conjunctivitis, and coryza (the last three being the

"three C's" of rubeola. During this time but usually prior to the rash, the characteristic Koplik spots, white to bluish spots on an erythematous base, may be found on the buccal mucosa. Usually they are fading or absent by the time of apparent cutaneous eruption.

The rash itself begins on the head and neck and moves downward. The rash forms over three days during which time the patient is the sickest. The rash fades in the same order it appears, often appearing brownish or coppery. While the leg lesions usually stay individual and discrete, the facial lesions will usually become confluent.

Atypical measles occurs in those who received the killed vaccine (pre 1963) and are later infected with the live virus. This usually has a more abrupt onset and a rash that starts on the extremities and spreads centrally. It may also be hemorrhagic or papulovesicular. Patients so affected may be quite ill.

Complications of measles includes pneumonia (either primary viral or secondary bacterial), encephalitis, exacerbation of tuberculosis, and the late complication of subacute sclerosing panencephalopathy.

Most diagnoses will be suspected clinically, especially if pathognomonic findings such as Koplik's spots are seen. Culture, from blood, urine, or pharynx, is possible but difficult. Complement fixation, hemagglutination-inhibition, direct immunofluorescence, or ELISA techniques may make more conclusive diagnosis. Acute and convalescent titers may be necessary in questionable cases.

Treatment is generally supportive. Vitamin A (200,000 IU for 3 doses) may be useful, especially in vitamin deficient populations, such as those seen during humanitarian missions abroad. Measles specific immune globulin may be recommended for household contacts of measles patients, especially for infants under 1 year-old, immunocompromised patients, and pregnant women. In the deployed setting, this is considered the primary therapy. Secondary infection should be watched for and treated as appropriate. (Prophylactic antibiotics are NOT recommended. The most effective treatment is prevention through appropriate childhood immunizations. The World Health Organization has targeted measles as a disease capable of eradication.

RUBELLA
(German Measles)

Infective from the end of the incubation period until clearance of the rash, Rubella is the only member of the *Rubivirus* genus and humans are the only known hosts. Epidemics usually occur in the spring and generally in urban population centers and in un-immunized victims.

Rubella , an RNA virus, has a 14 to 21 day incubation period and is shed in respiratory secretions. The first trimester of pregnancy is the most vulnerable time for intrauterine infection and development of neonatal rubella syndrome.

The prodrome is characterized by symptoms of a mild URI; low grade fever, headache, conjunctivitis, and lymphadenopathy may also occur during the prodrome. Flushing, macules, and papules begin on the face and rapidly migrate. As it appears on the trunk on the second day, it may already be clearing from the face. This helps distinguish rubella from rubeola that runs a longer course. The lymphadenopathy may be severe and cervical, occipital and postauricular nodes are most involved. Pruritus and mild desquamation may follow although usually not to the degree seen in conditions such as scarlatina or Kawasaki's disease. Except for occasional thrombocytopenia, complications are rare.

Cell culture is possible but tedious and variable in accuracy. Hemagglutination-inhibition for anti-rubella antibodies is the standard screening test although indirect ELISA tests are the most popular. IgM is used to diagnose intrauterine infections. IF and other antibody/antigen assays may also be employed. Most cases will be diagnosed on clinical grounds. Neonatal rubella has a variety of malformations including cardiac, ocular, auditory, orthopedic, central nervous system and hematologic.

Like measles, the best treatment is prevention through appropriate childhood immunizations. Treatment is supportive otherwise.

ROSEOLA
(Exanthem subitum, Sixth Disease)

Sero-positivity is fairly universal in the adult population and most infants are born with maternal antibodies. Active infection usually occurs between the ages of 6 months and 2 years of life, corresponding no doubt to waning of this natal protection and the constant exposure. The virus likely remains a latent infection indefinitely. This is caused by human herpes virus 6 and has an incubation of 5 to 15 days. The virus is shed in all secretions and likely spread by respiratory route.

The rash is proceeded by a characteristic prodrome of 3 to 4 days of very high fever in a child who is otherwise doing well. The fever onset may be quite abrupt and is a significant etiology of febrile seizures. Non-pruritic pink macules occur on the fourth day at which time the fever usually resolves suddenly. The macules blanch with pressure and often have a white halo surrounding them. Complications such as thrombocytopenia are rare. Atypical monocytes and a transient neutropenia may occur which are generally of no consequence.

Diagnosis is made on clinical grounds of the characteristic febrile prodrome followed by defervescence

and onset of rash.

As the rash is usually asymptomatic and the fever resolves with rash onset, no treatment other than reassurance to the parents is necessary.

FIFTH DISEASE (ERYTHEMA INFECTIOUSUM)

Occurring worldwide, it can affect all ages although school age children (5 to 15 years old) are those most likely affected. The incubation period is usually one to two weeks during which time the virus is being actively shed in respiratory secretions. Blood transmission can occur and is a factor in fetal infection

Parvovirus B19 is the cause of erythema infectiosum. This is not a veterinary disease and the parvovirus for which dogs are immunized is not infectious to humans. The blood group P-antigen is the viral receptor and those without this antigen cannot be infected.

Usually the first presentation is a macular eruption on face giving the characteristic "slapped cheek" appearance. A macular eruption over the extremities follows over the next week, usually most noticeable on the extensor surface. A reticulated erythema follows in the areas of prior involvement (cheeks, arms) which will fade only to dramatically worsen with heat, sunlight, or embarrassment. This is especially distressing to adolescents who may suddenly appear to erupt in a lace-like pattern on their cheeks weeks or even months after initial infection.

Papular-purpuric gloves and socks syndrome has recently been associated with Parvovirus B19. Presentation is, as described by the name of the syndrome, a purpuric eruption of the distal extremities with a very sharp cut-off of the eruption just proximal to the wrists and ankles.

Those with blood cell dyscrasias and other hematologic abnormalities are more susceptible to the unusual but severe complications of erythema infectiosum, these being aplastic crisis and hemolytic anemia. This is also the concern in intra-uterine infections. A serious, prolonged anemia may also occur in immune-compromised patients.

Diagnosis is usually made clinically. Culture is not generally available and antibody testing is only performed in a few research laboratories.

Treatment is supportive. Pregnant women who may have been exposed may have serologic testing (usually arranged through local health departments) and serial sonography. There is no available vaccine at this time. Immune-suppressed patients and those with aplastic crisis generally have higher viral shedding and should be placed in respiratory and contact isolation if hospitalized.

GIANOTTI-CROSTI DISEASE

Originally described as a papulovesicular eruption associated with Hepatitis B infection, we now recognize there are a number of different presentations of Gianotti-Crosti disease (GCD) and that it is associated with many different viral infections. It is also known by the descriptive terms of "papular acrodermatitis of childhood" and "papulovesicular acrolocated syndrome"

This occurs world wide with seasonal or clustered appearances coinciding to the underlying viral etiology. Children between the ages of 1 and 6 are most often affected.

An immune response to a variety of viral antigens, GCD has been associated with Human Herpes Viruses such as Epstein Barr virus, cytomegalovirus, HHV-6, Enteroviruses, respiratory syncytial virus, parainfluenza virus, parvovirus B19, and immunizations to MMR, polio, and influenza vaccines. Group A Beta-hemolytic streptococcus has also been associated with this eruption.

A viral prodrome occurs in many followed by a pruritic eruption which may be associated with malaise and lymphadenopathy. Symmetrical pink, flesh colored papules occur suddenly over the extremities, buttocks and face. Initially they may have a vesicular or eczematous appearance

Over time, several weeks, they will become more lichenoid. Symptoms following an uncomplicated viral infection, will usually resolve in 2 to 8 weeks.

Unilateral thoracic exanthem (asymmetric peri-flexural exanthem of childhood) may appear similar but is mostly confined to one side of the body. Others argue that this eruption is better considered a variation of GCD. Usually GCD spares the trunk. Early in the course and depending upon the presentation, contact dermatitis and drug eruptions might be considered.

Diagnosis of GCD is made clinically. There are no specific lab findings. In the right clinical setting CBC, throat culture, monospot, liver function tests or tests for hepatitis might be indicated.

Unless specific treatment is indicated because of an etiology such as streptococcus, treatment is supportive with consideration of topical anti-pruritics and oral antihistamines. Topical steroids are generally not effective.

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Bipolar Disorder in Children – A Real Entity or the New Fashion Diagnosis?

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If we thought that we were scratching our heads regarding accurate diagnosis of attention-deficit disorder (ADHD), oppositional-defiant disorder (ODD), and childhood depression, look out – now it's bipolar disorder. As parents come in distressed over their children with overactive, aggressive behavior, difficulties in school behavior, tantrums, and anxiety, there is increasing pressure in the lay medical literature pushing parents to find a new explanation. Parents, by the time they bring their child in to the primary care practitioner, are beleaguered with the responses from teachers, school administrators, other parents, and family members as to the wayward behaviors of their child. They are more likely now to want an explanation which certifies that their child is "ill" with a true disease (e.g., bipolar disorder), rather than having conditions such as ADHD and/or ODD, which in their minds carry a perception of being "bad". There is the implication that if the child is afflicted with a more "medical" condition, treatment will be more dependent on the practitioner finding the right medication in the office, rather than dealing with ongoing behavioral interventions at home and school.

So, what is the status of bipolar disorder in children – is it really an entity? The literature that parents are exposed to tells them that the professional community is too quick to make a diagnosis of ADHD, and that if a child with bipolar disorder is prescribed a stimulant, dire consequences will occur in the child with bipolar disorder. It is important to be able to assure parents that their child will receive a thorough evaluation, and that reasonable and helpful recommendations will be made as to treatment options. In order to feel comfortable making that happen, the provider needs to feel comfortable asking the right questions to differentiate between the "horses" and "zebras".

Bipolar disorder, as defined in its classical sense in the Psychiatric Diagnostic Statistical Manual (DSM-IV), is easier to recognize in adults and even adolescents, than it is in pre-adolescent children. Without minimizing the tricky nature of interpreting history and findings on the mental status exam, it is easier to recognize in adults intense depression, sadness, circumscribed manic episodes, hyper-religiosity, grandiosity, hyper-sexuality, pressured speech, sometimes irritable or euphoric affect, and evidence or complaint of racing thoughts Manic behavior in adults and adolescents is more circumscribed, whereas children have more chronic and less well define behaviors. Looking retrospectively at adults with bipolar disorder, one frequently sees that these individuals had childhoods that were characterized by behaviors which may have been interpreted as hyperactive, difficulties with focus, intense oppositionalness and tantrums, anxiety, and sadness.

The literature at this point is clear that, indeed, there is a case for the diagnosis of bipolar disorder in children, and that in fact, its early treatment has a significant impact on the psychosocial development of the patient. However, it has many overlapping behaviors with ADHD and ODD. Consider the following indicators of bipolar disorder in children:

(From *The Bipolar Child*, Demitri Papolos, 1999):

<u>Very Common</u>

- Separation anxiety
- Rages and explosive temper tantrums lasting up to several hours
- Marked irritability
- Oppositional behavior
- Rapid cycling (frequent mood swings, occurring within an hour, a day, or several days) or mood
- Lability
- Distractibility
- Hyperactivity
- Impulsivity
- Restlessness/fidgetiness
- Silliness, giddiness, goofiness
- Racing thoughts
- Aggressive behavior
- Grandiosity
- Carbohydrate cravings
- Risk-taking behaviors
- Depressed mood
- Lethargy
- Low self-esteem
- Difficulty getting up in the morning
- Social anxiety
- Oversensitivity to emotional or environmental triggers

<u>Common</u>

- Bedwetting (especially in boys)
- Night terrors
- Rapid or pressured speech
- Excessive daydreaming

Obsessional behavior
Compulsive behavior
Motor and vocal tics
Learning disabilities
Poor short-term memory
Lack of organization
Fascination with gore or morbid topics
Hypersexuality
Manipulative behavior
Extremely bossy behavior with friends/bullying
Lying
Suicidal thoughts
Destruction of property
Paranoia
Hallucinations and delusions

Less Common

Migraine headaches
Bingeing
Self-mutilating behaviors
Cruelty to animals

As is evident in the above-described behaviors, most of these behaviors can be seen in ADHD, ODD, conduct-disordered, or even otherwise normal children. In addition, whether or not these behaviors were actually abnormal would be dependent on the developmental level of the child in question. For instance, lack of organization would not be as much of concern until a child was close to middle school. We do not expect a 6-year-old to be particularly well organized. Another example is that destruction of property, manipulative behavior, cruelty to animals, and grandiosity are seen in conduct disorder. Oppositional behavior, distractibility, hyperactivity, impulsivity, restlessness/fidgetiness, silliness, giddiness, goofiness, and risk-taking behaviors are prominent in ADHD.

In considering these behaviors, there are two discriminating actions that are helpful in considering bipolar disorder in children. The first is to observe for clustering of several of these behaviors, to include other behaviors not as common in ADHD, ODD, or conduct disorder. The second would be to note an increase in severity or in intensity of the behaviors occurring in the more common disorders.

Thus, a child who had the behaviors usually associated with ADHD, but in addition, had a cluster of findings such as explosive tantrums for hours, racing thoughts, and rapid cycling, would cause the practitioner to consider the possibility of bipolar disorder. In another example, the oppositionalness and irritability frequently described in children who are frustrated with their ADHD, may be noted to be much more intense than expected. For example, irritability and defensiveness may be described by the parent as frequently exploding into violent behavior, destruction of property, harm and dangerous threats to others. This level of severity over and above what one usually sees in ODD/ADHD child would prompt a consideration of bipolar disorder.

Add to these histories episodes of depression, severe episodes of anxiety, and a family history of either depressive disorders, bipolar disorder, or substance abuse (frequently indicates self-medication for an a depressive disorder), and the index of suspicion should rise further. Any family history indicating some psychiatric disturbance like schizophrenia, “nervous breakdown”, “craziness”, “always depressed”, or “had to be in the hospital”, may indicate evidence of bipolar disorder which would not have been recognized at that time.

Psychological testing is less helpful or diagnostic, the younger the child. However, as the child gets older, testing, in conjunction with a careful history, can help to corroborate suspicions raised by history and mental status exam. Mania/depression on the Millon Adolescent Personality Inventory (MAPI) are examples of scales which may “light up” on psychological testing.

The main stays of treatment of bipolar disorder are the mood stabilizers. The most studied and still the most frequently used of these is lithium. It has its problems with fine tremors, and potential and/or transient GI, renal, thyroid, and neurological side effects, but these side effects are uncommon enough to make the use of lithium an appropriate choice for treatment. Other frquently used mood stabilizers are the anti-epileptics such as valproate, gabapentin (Neurontin), topiramate (Topamax), and lamotrigine (Lamictal). In addition, if manic episodes include psychotic symptoms such as delusions, hallucinations, or other evidence of disorganized thinking, neuroleptics (antipsychotics) such as risperidone and olanzapine can be effectively combined with mood stabilizers. If depression is a prominent feature of the bipolar disorder, the judicious use of antidepressant medications is helpful. There is the concern that the use of antidepressant medications will push the patient into a manic episode, especially if mood stabilizers have not been started.

While the diagnosis and treatment of children and adolescents with bipolar disorder will likely ultimately end up in the child psychiatrist’s office, it is important for the pediatrician to be aware of the possibility of this diagnosis. In addition, it may be approriate, once the symptoms have been stabilized, for the child’s primary care provider to manage medications if desired. The caveat is that environmental and other family stressors can frequently destabilize the child’s clinical status, and there should be consultation with a child psychiatrist in trying to adjust the medications. In addition, there needs to be associated psychotherapy, family therapy, and

close school consultation in order for the effects of the medications to be optimized.

It is not easy to diagnose bipolar disorder, especially in young children. However, it is appropriate to include it in the differential when ADHD or ODD appear to become increasingly complicated by their severity or accompaniment by more unusual behaviors.

Finally, while evidence is clear that bipolar disorder in children is real, it is still relatively rare. In addition, the implications of making the diagnosis are profound in labeling a child. It is a severe disorder, and its diagnosis results in the use of medications that carry potentially significant side effects and long term effects. Remember that ADHD and ODD and associated depression are far more common. The decision on a diagnosis of bipolar disorder should come only after careful observation, multiple episodes of history-taking, and possibly psychological testing. Ideally, consultation with or referral to a child psychiatrist is indicated.

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provide a brief reference for those who provide primary care for pediatric patients in our military health care system that will enable them to properly care for constipated infants and children without the need for sub-specialty referral. A much more detailed discussion of constipation may be found by reviewing the North American Society for Pediatric Gastroenterology, Hepatology, and Nutrition's Position Paper on Constipation published in November 1999.

Functional constipation can be defined most simply as constipation without any objective evidence of an etiologic, pathologic condition. The more confounding question is just what defines constipation. Attempts at defining constipation based on a single criterion such as stool frequency or stool consistency are notoriously unhelpful as such definitions may apply to a number of asymptomatic individuals with infrequent or firmer than usual stools. Furthermore, there may be a number of infants and children that have symptoms due to defecation problems or fecal retention that have daily stools that are not rock hard in consistency. I much prefer to think of constipation as a state in which one's stool consistency and/or their frequency of evacuation is insufficient to keep them free of symptoms. Clinical symptoms are the key to this working definition of constipation, but stool frequency and consistency clearly may be factors.

In terms of symptoms, these can be quite varied. No provider should have any difficulty in recognizing functional constipation in a child with infrequent, large, hard stools that are painful to pass. It requires a higher level of suspicion, however, to diligently rule out functional constipation as the etiology of so much of the non-specific abdominal pain that primary care providers and pediatric gastroenterologists also see, as patients and parents will not commonly report a history of constipation due to their own thoughts as to how this is defined. Thus, simply asking if the child is constipated frequently yields a negative answer, yet does not adequately cover the subject. The provider must diligently ask about the frequency, consistency, and size of the stools and about associated abdominal or anal pain or bleeding with defecation. Pain due to fecal impaction is typically periumbilical and crampy in nature. It is precipitated by eating via the gastrocolic reflex and is relieved, at least in part, by defecation. Patients may have overflow incontinence presenting as fecal soiling or staining in the underwear or they may not eat well when significantly impacted. Although nausea or vomiting should never be routinely considered as presenting symptoms of constipation, several children that I have cared for have had surprising resolution of such symptoms when their constipation was adequately managed, at least suggesting that such symptoms may be due to constipation in some cases.

When gathering the history, one of the most important pieces of information that can be obtained is

the timing of the onset of the problem. When asked when their child's problem began, many parents will simply say, "He's always been constipated" or "She's been constipated since she was a baby". It is vitally important to clarify *exactly* when the problem began, as this is crucial information when trying to differentiate between Hirschsprung's disease and functional constipation. Hirschsprung's disease, being a congenital absence of ganglion cells in the colonic wall that prevents appropriate relaxation and compliance in the affected colonic segment, inherently presents with problems in the neonatal period. As such, the passage of meconium after 48 hours of life should raise one's concerns for this diagnosis. In patients with functional constipation, however, there is some period of normal defecation that precedes the onset of constipation. Furthermore, the onset of functional constipation often coincides with dietary changes, life changes (e.g., PCS moves, divorces, etc.), or toilet training. Dietary changes that are notorious culprits for precipitating a pattern of constipation are weaning from breast milk to formula or whole milk, changing to soy formulas, the introduction of solid foods, and weaning from formula to whole milk.

The concern for an organic cause of the constipation is frequently the nature of the consult to the pediatric gastroenterologist. There are a number of organic conditions that can present with constipation, but a few screening questions in addition to a thorough physical exam should generally be able to dismiss these concerns. Patients with Hirschsprung's disease may present with constipation (or diarrhea), abdominal distension, and vomiting. They do not, however, have large caliber stools or fecal soiling except in very rare cases. A history of normal linear growth and development should allay concerns about hypothyroidism.

A thorough physical examination is critical in the evaluation of the constipated patient. A rectal exam is a must yet is the most frequently omitted part of the patient's evaluation prior to referral to the pediatric gastroenterologist. The rectal exam includes a measurement of the anogenital index to rule out anterior displacement of the anus, demonstration of a normal anal wink, screening for anal fissures and skin tags, and determination of the presence or absence of an anal stricture or stenosis, the size and compliance of the rectal vault, and the amount and consistency of any retained feces. Omitting the rectal exam is akin to diagnosing otitis media without looking in a patient's ears and is unacceptable. The physical exam should also include a thorough exam of the abdomen and the lumbosacral region as well.

An abdominal plain film radiograph should be ordered if one cannot demonstrate fecal retention by rectal examination, but the defecation history or history of abdominal pain are suspicious for functional constipation. I would caution the ordering provider, however, to review these films themselves as the degree of fecal retention is frequently not commented on by the radiologist, even when ordered specifically for this purpose. Obviously, the plain film radiograph may also reveal lumbosacral abnormalities that cannot be appreciated by physical examination.

Once the diagnosis of functional constipation is made, I begin by demystifying the disorder and sharing with them how exceedingly common it is in the pediatric population. It is very important to also discuss the pathophysiology of this disorder with the patient and the family as this will greatly enhance their compliance with treatment and make them a partner in the care of the problem. The colon acts as the trash compactor of the body and concentrates the waste products of small intestinal digestion and absorption mainly by absorbing excess fluids and electrolytes. Unfortunately, the colon can become too efficient in this role and can produce thick, pasty, hard stools in the process. The longer waste remains in the colon, the harder and drier it will become. The colon can also accommodate and enlarge to store feces until they are defecated. The longer the colon remains in this dilated state, the more compliance it develops and the more likely it is to remain dilated even when fully evacuated. A dilated colon with increased compliance will thus easily become reimpacted with feces.

Complicating matters further is the issue of stool withholding. Large, hard stools are painful to pass and may result in painful anal fissures. The reaction of the toilet-trained child who experiences such painful bowel movements is often to withhold stools so as not to have these painful bowel movements. After some time of withholding, the rectum accommodates, and the patient no longer experiences the sensation of having to defecate. Obviously, this can only last so long, and eventually the patient can no longer withhold the feces, which are now even larger and harder than before. An even more painful bowel movement occurs, and the response is more diligent stool withholding. Thus, a cycle is set up resulting in fecal impaction of varying degrees. The patient has accommodated to having a large mass of feces in the rectum and loses the normal sensation of when defecation should occur. If softer more liquid stool descends around the rectal impaction, this softer stool will frequently leak out in the form of defecation accidents and will only be detected by the patient when it stimulates the cutaneous nerves supplying the skin of the perineum or the olfactory senses of the patient or others

around them.

The most important points to make to the parents when explaining this pathophysiology are to emphasize that the encopretic patient truly cannot detect when he has to go to the bathroom nor can he sense when he's about to have an accident until it's too late. In essence, the encopretic child does not have voluntary control over when, where, or how he will go to the bathroom. It is critical that the parents understand this, as failure to do so can severely worsen an already troubling situation. It is important that the parents instead provide support and positive feedback to the encopretic child, especially as he makes progress in the management of the disorder. It is also important that the family understands that, just as it took many months for their child's problems to develop, it will also take a long period of time for the problem to resolve.

Now that we have discussed the pathophysiology of functional constipation, let's move on to the topic of management. Frequently, primary care providers attempt to manage constipation merely by managing diet. In general, when constipation has reached the point at which the family brings the child to a medical professional, the constipation is generally beyond management in this manner. Dietary alterations such as increasing fluid intake and increasing the amount of fiber in the diet may, indeed, be helpful at a later point in the management when attempting to wean the child off of stool softeners but are usually insufficient in treating a child with significant fecal impaction. Limiting whole milk intake, however, to 16-24 ounces per day may be somewhat helpful in decreasing problems with constipation but should always be balanced against the child's growth and nutritional needs. In general, once the constipated child presents to a primary care practitioner or a pediatric gastroenterologist, initial medical management will be necessary. The hallmark of therapy when treating a constipated child is to first cleanout the patient to relieve the fecal impaction and then to maintain a normal stooling pattern, so that over time the large intestine may return to normal size, compliance, and function. It is important that the cleanout occur and that it is complete. It is equally important that the maintenance therapy be sufficient to maintain a stool consistency and frequency that prevents fecal impaction from re-occurring. Again, it is important to spend additional time with the parents and the patient to emphasize the goals of both the cleanout and the maintenance therapy to assure compliance. Simply telling the family what medicines to take but without explanation of their intended effects or the duration of therapy will virtually guarantee noncompliance.

To demonstrate these above points, I would like to propose an example of a management strategy for treating chronic constipation. When I manage chronic constipation, I prefer to do oral cleanouts. Although a rectal cleanout for constipation may be just as effective when done properly, it may be a difficult regimen to comply with to the point of effectiveness, and it may further exacerbate the patient's fear of stooling and stool withholding. My preferred regimen for oral cleanouts is either magnesium citrate in a dose of 1 ounce of magnesium citrate per year of age (to a maximum of 10 ounces) per day for 5 to 7 days for children who cannot swallow pills or bisacodyl 5 mg tablets in a dose of 1 tablet by mouth per day for children < 10 years old or two tablets by mouth per day for children > 10 years old. After the cleanout, it is very important to bring the child back to determined if the cleanout was adequate. This is accomplished by taking a detailed history, performing a physical exam to include a rectal exam, and performing an abdominal radiograph if the history and physical are not adequate to make a determination as to whether the cleanout was effective or not. If the patient has been adequately cleaned out, then the maintenance phase of therapy is begun. If the child has not been adequately cleaned out, then the cleanout regimen is continued for approximately 3 to 5 days more, and the patient is again reassessed to determine if the cleanout has now been effective. In any case, once the cleanout has been adequately performed, the maintenance regimen is begun.

For maintenance therapy, I prefer to use magnesium hydroxide in a dose of one ml per kilogram of body weight per dose twice a day or two ml per kilogram of body weight once a day. Lactulose or mineral oil in the same dose are acceptable substitutes and may be better tolerated orally. I prefer to use magnesium hydroxide, however, as lactulose may cause more cramping due to the fermentation of the poorly absorbed carbohydrate, and mineral oil tends to result in more leakage accidents in between individual bowel movements. Magnesium hydroxide preparations come in many different flavors and, like the other stool softeners, may be mixed with virtually any sort of food product in an effort to mask the taste and to achieve patient compliance. For those patients who still cannot tolerate magnesium hydroxide preparations, I will switch them to polyethylene glycol without electrolytes in a dose of 17 grams in eight ounces of water for children > 5 years old and half that amount for children < 5. The advantage to the polyethylene glycol preparation is that it is tasteless. Unfortunately, the polyethylene glycol preparation requires a prescription and is rather expensive. The other stool softener options are available as over-the-counter medications and are

relatively inexpensive.

Regardless of the stool softener chosen, the goal of therapy remains the same. The dose should be titrated upward or downward to achieve at least one, soft, easy-to-pass, non-formed bowel movement per day. It is important that the patient have a bowel movement daily for the period following the cleanout so that fecal impaction will not recur. The ideal consistency of the stool during this period is roughly that of mashed potatoes, however, I usually err on the side of the stools being looser if that is what it takes to achieve a daily bowel movement frequency. The polyethylene glycol solution seems to be an exception to this case in that soft, but formed, stools seem to be an adequate consistency for maintaining the effects of the cleanout. The maintenance dose is then kept constant for a period of several months (average of 4-6) after which it is slowly weaned. Of critical importance in weaning process is ensuring that the patient continues to defecate normally as the dose of the medicine is being decreased. If the patient has a setback during the weaning period, the maintenance dose of stool softener should be increased to the last effective dose, and no further weaning should be attempted for several weeks.

The preceding example of the cleanout and maintenance regimen for treating chronic functional constipation provides the primary care practitioner with a framework for managing this problem. If these principles of management are followed to the endpoints described, the primary care practitioner should have no difficulty in managing the patient with chronic functional constipation. There are several pitfalls, however, that the primary care practitioner frequently encounters when evaluating a patient with functional constipation that seem to result in an unnecessary referral to pediatric gastroenterology. In the order in which they occur, these are as follows. First of all, the diagnosis of functional constipation is frequently not made. This is often due to failure to ask detailed questions about the patient's defecation history or failure to recognize an abdominal pain pattern consistent with chronic functional constipation. Omitting a rectal exam as part of the physical examination and/or failing to perform an abdominal radiograph as part of the evaluation further reduce the chances of correctly making this diagnosis. If, however, the diagnosis is correctly made, another common mistake made is failing to clean out the patient that presents with fecal impaction. If a cleanout is attempted, many practitioners do not bring the patient back for a follow-up examination immediately after the cleanout period to determine whether it was effective or not. Lastly, many practitioners will not use adequate doses of stool softeners or will allow the family to prematurely stop using them. This is often due to false assumptions that many providers have about the long-term use of stool softeners. Unlike stimulant laxatives, which are absorbed by the body and short circuit the body's normal method of having a bowel movement and upon which patients may develop a dependence, all of the above listed stool softeners work by not been absorbed. Given this mechanism of action, there truly is no "dose" of a stool softener, as one needs to use as much or as little as it takes to achieve the desired endpoint. Stool softeners merely adjust the consistency of the stool while allowing the patient to defecate in a normal physiologic fashion. Any method by which the patient could achieve the same stool consistency would be just as effective. Therefore, practitioners, families, and patients do not need to have the frequently expressed unreasonable fear of using stool softeners for a long period of time, as they do not create dependence.

In this brief discussion of the presentation, pathophysiology, and management of chronic functional constipation, I have attempted to present some of the most basic points about this entity that the primary care practitioner can use to effectively care for their patients affected by this disorder without having to refer them to the subspecialist. Clearly, those patients who have constipation due to an entity other than a functional one should be referred. I have also attempted to present some of the most common errors made in the evaluation and treatment of patients with chronic functional constipation to guide the provider away from the common mistakes that we see in the management of chronic functional constipation patients that are referred to pediatric gastroenterology. Hopefully, the combination of these discussion points will improve the ability of the primary care practitioner to care for patients affected with this disorder.

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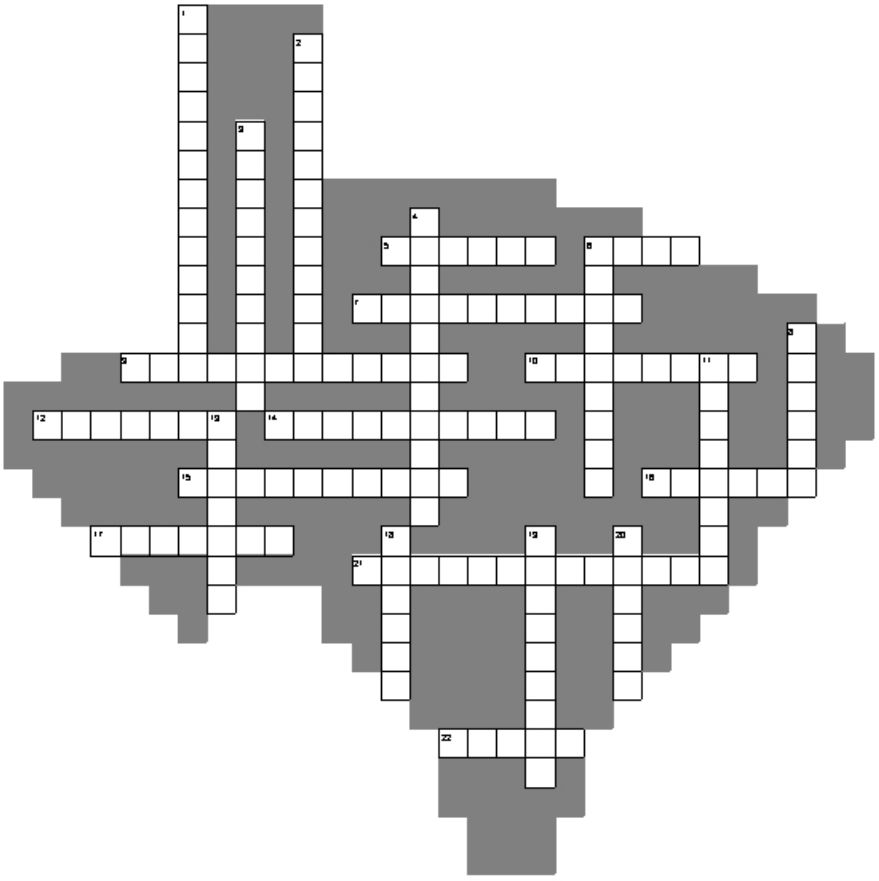
October 2002 Crossword Puzzle (3)



Word List

Actionplan
Aneurysm
Aspirin
Asthma
Asymptomatic
Circumcision
Controllers
Curtsy
Guidelines
Koplik
Luque
Maculopapular
Morbus
Nonstructural
Parvovirus
Pili
Platelets
Pyuria
Roseola
Rubivirus
Sagittal
Spirometry
Synergy

Across

5. Old name for plague
6. Enteropathogenic E coli adhere to epithelial cells via these filaments
7. An important written piece of asthma education
9. Meta analysis of multiple studies has shown that this procedure may decrease the risk of infant UTI by a factor of 12
10. Primary adverse sequela of Kawasaki syndrome
12. Sixth disease synonym
14. Instruments that help standardize disease management using evidence based medicine
15. The best objective evidence of airways obstruction
16. The most common chronic illness in childhood
17. When the bladder contracts in coordination with the bladder neck relaxation
21. A flexible scoliotic curve that compensated for a true scoliotic curve, allowing the head to remain balanced over the pelvis
22. Mexican surgeon who pioneered the instrumentaion of scoliosis using sublaminar wires at every level to fix rods to the posterior elements of the spine

Down

1. One type of rash seen in KAwasaki syndrome
2. The finding of bacteria in the urine without symptoms of infection
3. Fifth disease etiologic agent
4. Medications used on a daily basis to treat persistent asthma
6. Can often exceed 1 million/mul during the second or third week of illness in Kawasaki syndrome
8. One third of patients with Kawasaki syndrome have this urinary finding
11. A deformity in this plane of the back is a disruption in the normal thoracic kyphosis and lumbar lordosis that accompanies scoliosis
13. Used in the treatment of Kawasaki syndrome for its antiinflammatory and antiplatelet effects
18. Eponym for the buccal lesions of early measles
19. Etiologicagent of rubella
20. Vincent described this position when a child squats and places the heel of the foot in the perineumto help augment the external sphincter

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